

Case Report

An Newborn Patients with A Pentalogy of Fallot with Pulmonary Atresia with Mild Hydrocephalus Case

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Abstract

Tetralogy of Fallot (TOF) in conjunction possessing a visible Foramen Ovale (PFO) or an Atrial Septal Defect (ASD) characterises that uncommon cyanotic congenital heart condition known as Pentalogy of Fallot (POF). TOF is characterised by overriding aortic root, right ventricular hypertrophy, narrowing of the Outflow tract of the right ventricle, and Ventricular Septal Defect (VSD). The pentalogy of Fallot is a variation of the additional frequent tetralogy of Fallot and has the four standard characteristics plus an Approved ductus or atrial septal defect arteriosus. Ventricular Septal Defect (VSD), tightness of the right ventricular outflow tract or blockage right ventricular hypertrophy surpassing the aorta in size, or Patent Ductus Arteriosus (PDA).

A birth defect of the pulmonary valve known as pulmonary atresia regulates circulation coming from the right ventricle, which is the lower right chamber of the heart, to the major pulmonary artery, which is the blood vessel that carries blood from the heart to the lungs. There is no blood flow from the right ventricle of the heart out to the lungs in pulmonary atresia because this valve did not develop at all. An inherited heart condition is called pulmonary atresia Congenital Heart Disease (CHD) that is classified as a significant CHD because a newborn with it may require surgery or other operations soon after birth. Congenital denotes existing at birth. The blood vessel that carries blood from the heart to the lungs does not open and close, but instead develops into a solid sheet of tissue. Therefore, the blood's typical path to the lungs to absorb oxygen is blocked. An alternative route for some blood to reach the lungs is through the heart's and its arteries' natural ducts.

Keywords: Uncommon cyanotic congenital heart; Uncommon cyanotic congenital heart; Patent ductus arteriosus; Right ventricular hypertrophy overriding aorta

Introduction

A born baby below 28 days of age is known as a newborn or neonate. And the baby's greatest risk of death occurs in these first 28 days of existence. Most of the neonates die in developing countries due to the restricted access to healthcare, which indirectly leads to the increase in Neonatal mortality rate. In tetralogy sufferers to pentalogy of Fallot and Pulmonary Atresia (TOF/PA), the lung valve, which opens to permit blood to pass from the right ventricle to the lungs, is atretic, a severe variation of the illness [1,2]. Too little blood reaches the lungs in the majority of Tetralogy of Fallot cases. After that, the low-oxygen (blue) blood travels throughout the body, depriving the tissues of oxygen [3]. Tetralogy of Fallot (TOF), including Pulmonary Stenosis (PS), a One variation of the more popular Pentalogy of Fallot

(POF) protocol an Atrial Septal Defect (ASD), ventricular septal defect, right ventricular hypertrophy, and an overlapping aorta, and no epidemiological data is available. When an adult has congenital heart disease, asymptomatic POF is uncommon, particularly in emergency patients [4]. The Fallot's a tetra, or tetralogy of Fallot, is characterised by four main characteristics. The most significant are pulmonary stenosis, or multiple degrees of blockage from the right ventricle to the lungs, and ventricular septal defect, or a hole between the ventricles. Furthermore, the ventricular septal defect is directly beneath the aorta, the main artery that connects the heart to the body, while thicker muscle grows in the right ventricle. Because the aorta overrides the ventricular defect and there's pulmonary stenosis, the body receives blood from both ventricles-those with more oxygen and those with less. Pulmonary atresia is the condition in which the pulmonary valve is entirely blocked. As in this instance, babies and young children with unrepaired tetralogy of Fallot are frequently blue (cyanotic). The body pumps some blood that is low in oxygen [5]. A congenital heart defect known as pulmonary atresia is typically discovered shortly after birth. In pulmonary atresia, the valve that discharges blood into the lungs from the heart (pulmonary valve) is not properly formed. Rather than opening and shutting to let blood to travel from the heart to the lungs, Tissue congeals into a thick sheet. So, Blood cannot flow in the normal way to pick up oxygen from the lungs. As an alternative, some blood goes to the lungs through other natural passages throughout the arteries and heart.

During a baby's development in the womb, these passages

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are essential, and they typically near soon after birth. Babies with pulmonary atresia often exhibit a blue tint to their skin as a result of not being getting enough oxygen.

A life-threatening condition is pulmonary atresia. Treatments to improve a baby's heart health and drugs to make the baby's heart function better are the initial measures in the management of pulmonary atresia [6]. Definition of TOF could be based on of the outlet's anterocephalad deviation septum with septoparietal trabeculation-related malformation or, as an alternate, may result from the sub pulmonary infundibulum is still developing. A during embryogenesis, a single anomaly or pathological process results in Right Ventricular Outflow Tract (RVOT) hindrance as well as VSD of the malalignment kind and aortic override. Ventricular hypertrophy on the right is the hemodynamic outcome of the over physical lesions.

TOF exhibits a broad range of illness, RVOT obstruction varying in severity. In cases of TOF accompanied by pulmonary atresia, a severe blockage could happen at 1 or more of the following structures: (a) infundibulum, (b) pulmonary valve, (c) main pulmonary artery [7]. Hydrocephalus is a condition in which the Cerebrospinal Fluid (CSF) within the cavities of your brain and the spinal cord gets accumulated or gets deposited in one place. These empty spaces where they used to get deposited are called ventricles. The buildup of these CSF can apply a hard pressure to the brain [8]. The impact of hydrocephalus on a newborn is reasonably erratic and can vary in severity. As there was a pressure on the brain due to accumulation of CSF it causes many problems to the newborn like brain damage, epilepsy, learning impairments, short-term memory loss, and difficulties in coordinating, vision problems, and early onset of puberty.

As a result, children with this condition frequently gain from developing therapy, such as physical therapy and occupational therapy. In softer instances, or instances where treatment was formerly feasible, a baby with hydrocephalus may develop in a fully typical manner. We realise that cerebral hydrocephalus can be a scary diagnosis. That's what makes us available every day, seven days a week. Even though we are powerless to alter the diagnosis, we can still show compassion and support, you seek, helping your child get the most removed from care and existence [9].

Case Presentation

A 1.75 kg male child born to G4P2L1A1D1 mother with 38.4 weeks of gestational age via LSCS in view of cervical stitch insitu & Baby cried immediately after birth. Apgar at 1, 5 mins was 9, 10. Child was shifted to NICU for observation i/v/o oligodactyly, absent eyebrows, micrognathia. On admission in NICU child was given katori spoon feeding trail child was accepting and tolerating feed well the child was put on nasal prongs for O₂. Were done s/o b/l ventricles appear dilated with multiple septae within and advised MRI for further evaluation. MRI was done with impressions of ependymal Cysts in frontal horn of bilateral lateral ventricles, hypoplasia of posterior corpus callosum. 2d echo was done were impressions of pentalogy of fallots with pulmonary artesian was noted.

On physical examination, A 1.75 kg male child born to G4P2L1A1D1 mother with 38.4 weeks of gestational age via LSCS in view of cervical stitch insitu & Baby cried immediately after birth. A newborn with TOFPA frequently exhibits symptoms in their first few hours of life. The ducts start to close after birth, resulting in severe cyanosis. There could be mild to moderate cyanosis if there are significant acras pulmonary collaterals. If there are insufficient

collaterals to sustain enough pulmonary flow, the ductus arteriosus may close, potentially leading to fatal hypoxemia. Patients who exhibit ductus persistent patency or well-developed aorto-pulmonary collaterals occasionally present with respiratory compromise as a result of over circulation of the lungs and heart failure. As resistance of the lungs to oxygen falls and pulmonary blood flow rises in the weeks following delivery, symptoms start to appear. Babies may not gain weight if they eat poorly (Figure 1 and 2).

Discussion

The macroscopic lesions seen during necropsy, the ultrasonography results, and the clinical indicators all contributed to the pentalogy of Fallot diagnosis. One of the most prevalent congenital cardiac malformations in horses is tetralogy of Fallot; yet, in a study of inherited deformities in this species, only 3.5% of the horses had cardiac deformities out of the 608 foals that were inspected. In contrast to other domestic animals, horses have a comparatively low occurrence of cardiac abnormalities. Between 0.1% and 0.5%, since the foal was the sole animal to be born with the deformity in this instance, it was most likely sporadic. (10) The term "Pentalogy of Fallot" describes how an atrial septal defect and Tetralogy of Fallot (TOF). One At 3 per 10,000 live births, TOF is the most prevalent type of birth defects of the heart that are cyanotic, comprising between 5% and 10% of all Congenital Cardiac Conditions (CHDs). The aorta overrides the nonrestrictive perimembranous VSD, RV hypertrophy, RV outflow blockage, and antero-cephalad displacement of the infundibular septum are the characteristics that define it [11]. The POF is a variation on the more widely recognized Tetralogy of Fallot (TOF), which includes an atrial septal defect in addition to the four conventional characteristics. Despite being referenced by Niels Stensen and Edward Sandifort, it was not until 1888 that the physician Étienne-Louis Arthur of France Fallot-after whom it is named-made a thorough investigation and description of it [12]. In most patients with intact ventricular septum and pulmonary atresia, the right ventricle is a tiny chamber with solid walls. Two on radiography it is typically possible to see that a closed, obstructed right ventricle is the source of radiopaque dye entering the coronary vascular bed in more greater than half of patients in this category (type I)." The widely recognized explanation for this atypical communication is that the blood's unusually high pressure causes channels that are typically present in the growing heart to stay open [13]. The veins in the lungs in people with common pulmonary vein atresia are not connected to the left atrium. The esophageal vein or the bronchial venous system vein is where they empty into instead of joining together as a vein convergence near the rear portion of the left atrium [14]. The buildup of too much CSF inside the skull cavity, which causes an anomalous dilatation of the referred to as the subarachnoid space or ventricular system to as hydrocephalus [15]. Unified hydrocephalus in either the normal or disordered foetus can be detected prenatally, although this is challenging since typical sonography artefacts can mask foetal intracranial anatomy and, in certain situations, mimic the findings of unilateral hydrocephalus. In the dependent cerebral hemisphere, for instance, the anechoicity of the normally growing cortical mantle may frequently indicate unilateral ventricular enlargement if the lateral ventricular wall is not clearly defined [16]. However, sedation is often needed for newborns having MRIs, even though the new generation of CT scanners with their quick picture acquisition speeds usually avoids this need. This is a major benefit when cyanosis is present since sedation can worsen respiratory depression [17]. In our study, the overall mortality rate is 6.66%. In comparison to studies by Pan,

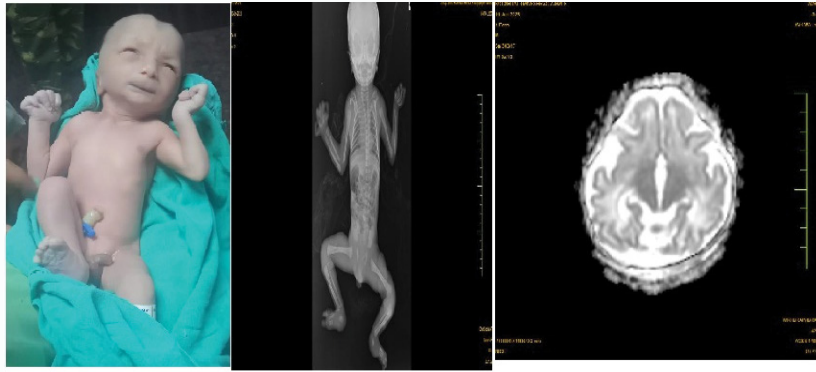


Figure 1: MRI Scan.

Patient Name	B/O SHITAL GANESHRAO AJBALE			Age & Sex	4 Days&M
Patient UHID	2300223833	Patient ID	2306100136	Study Date	14-Jun-2023
Patient Type	IPD	Modality	MR	Report Date & Time	14-Jun-2023 16:08:04
Consulting Dr.	DR. P. H. PARIHAR		Referred By	NICU	
Study Name	MRI BRAIN(BRAIN)				

CEMRI BRAIN

- There is e/of thin walled non enhancing cystic lesion appearing hypointense on T1WI/FLAIR, hyperintense on T2 noted in frontal horns of bilateral lateral ventricles.
- Posterior part of the body and splenium of corpus callosum appears thinned out (apple core appearance).
- There is e/o dilated cavum septum pellucidum measuring 1.7x0.6 cms.
- Brain parenchyma shows normal gray white matter differentiation.
- Both gangliocapsular region appears normal
- In the Infratentorium, the Brainstem, the Cerebellum and The Basal Cisterns appear normal.
- Sella and parasellar region appears normal.
- Visualized portions of the paranasal sinuses and orbit are normal.

IMPRESSION CEMRI BRAIN REVEALS:

- EPENDYMAL CYSTS IN FRONTAL HORN OF BILATERAL LATERAL VENTRICLES.
- HYPOPLASIA OF POSTERIOR CORPUS CALLOSUM.

ADVICE :- CLINICAL CORRELATION

TECHNICIAN – MR. BHOLE JR2- DR.SHEETAL JR1- DR.SHUBHI/DR.PASCHYANTI

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Date: 14-Jun-2023 16:08:04

This Report is Electronically Validated and does not require a signature

Figure 2: MRI Report.

the mortality rate is lower [7-9]. This can be as a result of our setup's Neonatal Intensive Care Unit (NICU) accessibility services for post-operative patient supervision, skilled neurosurgery facilities, and enhanced anesthetic techniques [18].

Conclusion

We discussed a patient's case that was identified as having TOF in his few days. The case suggests that despite advancements in medicine, individuals with primary congenital diseases might not receive a

diagnosis until they are older. There is a greater chance of negative consequences if TOF and pulmonary atresia cases are not diagnosed and treated promptly. But our patient's post-operative and two-day follow-up profiles were both very good. A screening echo in the early stages of life and a thorough physical examination of newborns may help identify the illness sooner.

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